

### 239 Heart assessment during exercise can predict pulmonary hypertension (PH) in CF

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**Background:** PH is recognized as predictor of severe prognosis in CF lung disease. Left ventricular (LV) systolic or diastolic dysfunction has also been reported.

**Aim:** to identify possible echocardiographic findings of cardiac dysfunction during both rest and exercise in adults with CF.

**Methods:** 24 patients (pts)(mean age 31.6, range 13–55 yrs; 14 males) with moderate-severe airway obstruction (mean FEV1 42, range 21–104% pred.) underwent to spirometry, conventional doppler echocardiography (DE) and tissue doppler echocardiography (TDE). TDE was performed at the level of lateral and medial mitral annulus (LMA, MMA), tricuspid annulus (TA), right ventricular (RV) free wall (FW). RV function was qualitatively assessed in 4 chamber view. Tricuspidal annular plane systolic excursion (TAPSE) was also measured. Echocardiography was performed during an incremental exercise test on an ergocycle in 13 pts with severe lung disease (FEV1 <35% pred.).

**Results:** Tricuspid regurgitation (TR) was detected in 4/24 (16%) patients, in 2 of them TR maximum velocity exceeded 3 m/sec. TAPSE (mean value 22.8, range 14–27), RV and LV systolic and diastolic parameters were within normal limits in all pts. Mean TDE values were normal at all levels (LMA 11.57 cm/s, MMA 8.64 cm/s, TA 9.22 cm/s, RVFW 8.3 cm/s). Dyskinesia of the ventricular septum (VS) but not TR was shown in 5/13 (38%) pts during exercise. None of these had PH findings using DE and TDE. Wall motion was normal at all the other levels.

**Conclusions:** Using DE and TDE we found no abnormality of systolic and diastolic RV and LV function and PH in only 2 pts. Using exercise echocardiography VS dyskinesia was found in about one third of pts with severe lung disease suggesting effort PH.

### 240 The autopsy reports: Cor pulmonale in CF patients with aging

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From definitions provided by the World Health Organization; cor pulmonale (pulmonary heart disease), pre-terminal and secondary disease in CF patients involve “hypertrophy of the right ventricle resulting from lung disease.” It is believed that cor pulmonale in CF is related to the development of lung destruction with progressive pulmonary hypertension. Although a large amount of study effort has been directed toward right atrium and ventricle dysfunction in CF patients, very few studies have been done for left ventricle dysfunction and coronary artery disease. We examined the 187 autopsy reports (77 males and 110 females) that were requested and performed with the permission of patients or their family from 1940s to 2000s with the age distribution from 5 days to 50 years. In evaluations of heart condition for the autopsied CF patients, 68% showed evidence of cor pulmonale (right side heart failure) and 20% showed evidence of problems in both ventricles. We grouped the autopsied CF patients into 4 groups; age at death under 10 years old, b) 10 to 19, c) 20 to 29 and d) over 30 years old to evaluate cardiac dysfunction with respect to age. Cor pulmonale was presented in 41% of the patients in group (a); 61% in group (b); 85% in group (c) and 85% in group (d). This result showed that the risk of cor pulmonale in CF patients increased as they aged. The occurrence and progression of heart failure in CF is individually and circumstantially different. There is no correlation between pulmonary function tests (PFT) and the appearance of heart disease in CF patients by observing PFT tracking.

### 241 Improvement in aerobic fitness and quality of life does not depend on severity of pulmonary disease in CF patients

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**Objective:** Exercise training programs to improve aerobic fitness are recognized as essential components of pulmonary rehabilitation except in respiratory insufficient individuals. The aim of this study was to determine, whether improvement of aerobic fitness (VO2peak) and quality of life (QoL) during an inpatient rehabilitation program depend on disease severity, gender or age in Cystic Fibrosis (CF) patients.

**Methods:** 79 CF subjects (38 females) with a median (quartiles) age of 22.9 years (15.8, 29.9) were analyzed for change of VO2peak and QoL before and after a 3-weeks inpatient rehabilitation program including daily physical exercise training. Severity of pulmonary disease was defined according to FEV1 at study entry (mild >60%, n=26; moderate 40–60%, n=26; severe <40% predicted, n=27). Influence of gender and age (<20 yrs, 20–30 yrs, >30 yrs) was investigated.

**Results:** Both VO2peak and QoL improved significantly during the rehabilitation program for the total population as well as for all subgroups (p < 0.05). Subjects with mild, moderate and severe lung disease improved VO2peak and QoL in the same manner (p=0.643 and p=0.283, respectively). No differences in improvement of VO2peak and QoL could be detected between gender (p=0.206 and p=0.060, respectively) and age groups (p=0.454 and p=0.907, respectively).

**Conclusion:** A 3-weeks multidisciplinary rehabilitation including daily exercise training improves aerobic fitness and QoL in cystic fibrosis patients independently of disease severity. Patients with severe pulmonary disease and respiratory insufficiency show no differences in trainability compared to mild and moderate affected subjects.

### 242\* Respiratory exacerbations in children aged 0–3 years in the Australasian Cystic Fibrosis Bronchoalveolar Lavage (ACFBAL) Study

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**Background:** Respiratory exacerbations in CF children causes progressive lung scarring. However, the ‘usual’ exacerbation rate in early years and the importance of individual clinical features observed is unknown.

**Aim:** To describe the frequency and clinical parameters associated with respiratory exacerbations in children with CF diagnosed on newborn screening and followed prospectively for 3 years.

**Methods:** All respiratory exacerbations were recorded. Clinical symptoms and signs, treatment with oral antibiotics, and admissions to hospital for investigation and/or intravenous antibiotics were documented.

**Results:** We enrolled 168 children at mean age 3.6mths (SD 1.6) from 8 Australasian sites, with 159 (81 boys, 78 girls) completing 3 years. 1132 exacerbations were recorded with 114 resulting in hospital admission. There was a median of 6 (range 0–20) respiratory exacerbations/child. The rate of exacerbations/year varied little (mean 2.56, 2.55, 2.59, SD 2.20, 1.93, 1.66) respectively. There was no difference in exacerbation rate between boys and girls P=0.55, or between children on (2.08 95% CI 1.72, 2.52) or not on (2.62 95%CI 2.46, 2.78) antibiotic prophylaxis with hazard ratio 0.87 (95%CI 0.69, 1.09 P=0.23). The most common features associated with exacerbations requiring oral antibiotic treatment was the presence of cough (94%), wheeze (28%), and crackles (4%).

**Conclusion:** Australasian children with CF have a rate of 2.6 respiratory exacerbations per year in the first 3 years with 90% treated in the community and 10% requiring hospitalisation.

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